

## Case report

### **A rare presentation of solitary pedunculated lipofibroma of the thigh: Case report and review of literature**

#### **ABSTRACT**

**Background:** Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartomatous idiopathic condition.

**Case Representation:** This case report describes a solitary and pedunculated lipofibroma, a rare variant of NLCS with review of the literature.

**Conclusion:** Early recognition of this benign condition will result in less morbidity amongst the patients with regards to cosmesis.

**Keywords:** Pedunculated lipofibroma, solitary, NLCS, lipoma

#### **INTRODUCTION**

Hoffman and Zuhrelle in 1921 first described a rare benign hamartomatous idiopathic condition called nevus lipomatosus cutaneous superficialis (NLCS).<sup>[1]</sup> It is characterized by the presence of ectopic mature adipose tissue within the dermis. Literature describes two clinical variants - the classical form, consisting of multiple skin-colored cerebriform, pedunculated nodules that most often combine to form a plaque and the second one which is relatively a rare form, presenting as a solitary sessile papule or nodule usually dome-shaped.<sup>[2,3]</sup> This case report describes a solitary and pedunculated lipofibroma, a rare variant of NLCS with review of the literature.

#### **CASE REPORT**

A young gentlewoman aged 27 years presented with a four-year-old history of a swelling on the inner side of left upper thigh. The swelling had originally appeared without any trigger and

gradually increased in size. No significant family history. Physical examination showed a pedunculated, solitary nodule measuring 40×40 mm. The prominent stalk was 10 mm long. The swelling was skin-colored, nodular in appearance, soft in consistency, non-tender, with no sinuses or ulceration. (Fig 1A) Surrounding skin appeared normal. No other swellings were noted. Fine needle aspiration of the swelling reported scattered mature adipocytes with many fibrous strands against a hemorrhagic background. Under local anesthesia, an elliptical incision was marked around the stalk and deepened to completely excise the swelling. (Fig 1B) Incision was closed using non-absorbable sutures (polyethelene 4-0). Histopathological examination (HPE) of the swelling showed infiltration of the epidermis and dermis with chronic inflammatory infiltrate. Mature adipocytes were seen in lobules separated by fibrocollagenous stroma. Blood vessels were also seen in the stroma. No malignancy noted. (Fig 2A,2B,2C) The findings were suggestive of NLCS, solitary pedunculated variant. Post-operative period was uneventful. Sutures were removed in two weeks after the surgery. One year follow-up showed no recurrence.

## **DISCUSSION**

Mehregan et al., suggested the term “pedunculated lipofibroma” for the solitary form of nevus lipomatosus, in view of its characteristic clinicopathological features.<sup>[4]</sup> Pedunculated lipofibroma is a rare, benign connective tissue neoplasm. It is solitary, slow growing and is distinctively diagnosed by the presence of ectopic adipose tissue in the dermis. The histological features are like NLCS, which was originally classified into two clinical types, a multiple form, and a solitary form<sup>[1]</sup> In the multiple type of NLCS, the lesions are either congenital or develop during the first three decades of life. The common sites of occurrence are usually the gluteal region, lower back, and the superior, posterior thighs. The solitary nodular, papular or pedunculated lipofibroma occurs later in life (usually over 20 years of age) and is found in unusual sites like axilla, knee, eyelid, nose, ear/pinna, clitoris, scrotum, or skin of the scalp.<sup>{5,12,13}</sup> NLCS are slow growing and asymptomatic in nature. However, large lesions tend to ulcerate with superimposing bacterial infections.<sup>[2]</sup> Ulnar neuropathy due to compression by a large NLCS has been reported where a partial excision of the lesion was done due to its benign nature.<sup>{9}</sup> There is no documented genetic predilection, but one study has reported the association of 2p24 deletion.<sup>{10}</sup>

The pathogenesis of pedunculated lipofibroma is unknown. Various theories have been postulated vis-a-vis degenerative changes in the collagen and elastic tissue resulting in the displacement of subcutaneous adipose tissue into the dermis. Another theory explains origination and differentiation of lipoblasts to adipose cells from the walls of dermal capillary vessels.<sup>[5,8]</sup>

The main histological abnormality in either type of NLCS is ectopic fatty tissue in the upper dermis, distinct from the normal subcutaneous fat. In many cases, the connective tissue has been seen to be organized irregularly. Foci of adipocytes are seen around the dermal blood vessels.<sup>[6]</sup> Staining with alcian blue shows increased deposition of mucopolysaccharides in the reticular dermis and fatty tissue in majority of the cases.<sup>[7]</sup>

Clinically, the differential diagnosis for a pedunculated lipofibroma includes other benign papillomas, including acrochordons, seborrheic keratosis, nevocellular nevi, verrucae, neurofibromas, fibroepithelioma of Pinkus, and eccrine poroma.<sup>[5]</sup> HPE will form the key contributor to the final diagnosis.

The treatment of pedunculated neurofibroma involves complete excision for both therapeutic and cosmetic reasons. Excision of large sessile plaques may result in the need for skin grafts or a combination of tissue expansion and flap advancement surgery to cover the post-excision defects. Other non-surgical modalities such as cryotherapy, carbon dioxide ablative laser, corticosteroids topical applications and intralesional phosphatidylcholine and sodium deoxycholate injections have been used for the removal of these lesions.<sup>[11,14,15]</sup> Recurrences have been noted following these modalities when surgical excision becomes an absolute necessity.<sup>[14]</sup> Although there are no reports in the literature about any malignant transformation of pedunculated lipofibroma, it is prudent to examine these swellings for malignancy considering the incidence being rare.

## **CONCLUSION**

NLCS is a rare skin pathology and pedunculated lipofibroma appears to be a rarer variant. Early recognition of this benign condition will result in less morbidity amongst the patients with regards to cosmesis. The comprehensive review of the literature has suggested that surgical excision remains to be the gold standard treatment to prevent recurrence and early intervention will prevent the need for large reconstructive options unless indicated.

## **STATEMENTS AND DECLARATIONS**

Informed consent

Informed consent was obtained from the patient for the use of photographs for publication. The consent has been formally documented in the medical record.

## **REFERENCES**

1. Hoffmann E, Zurhelle E. Über einen naevus lipomatodes cutaneus superficialis der linken Glutäalgegend. Arch Dermatol Syphilol 1921; 130: 327–333.
2. Ghosh SK, Bandyopadhyay D, Jamadar NS. Nevus lipomatosus cutaneus superficialis: an unusual presentation. Dermatol Online J. 2010;16:12.
3. Al-Mutairi N, Joshi A, Nour-Eldin O. Nevus lipomatosus cutaneus superficialis of Hoffmann- Zuhrelle with angiokeratoma of Fordyce. Acta Derm Venereol. 2006;86:92-93.
4. Mehregan A.H., Tavafolghi V., Ghandchi A. Nevus lipomatosus cutaneus superficialis (Hoffmann-Zuhrelle) J Cutan Pathol. 1975;2:307–313. doi: 10.1111/j.1600-0560.1975.tb00183.x.
5. Kwon KS, Seo KH, Jang HS, Chung TA. A case of apple-shaped pedunculated lipofibroma. J Dermatol. 1997 Apr;24(4):258-61. doi: 10.1111/j.1346-8138.1997.tb02785.x.
6. Öztürkcan S, Terzioglu A, Akyol M, Altınor S, Yıldız E. Pedunculated lipofibroma. J Dermatol 2000; 27: 288–290.
7. Nogita T, Wong TY, Hiden A, Mihm MC, Kawashima. Pedunculated lipofibroma. A clinicopathologic study of thirty-two cases supporting a simplified nomenclature. J Am Acad Dermatol 1994; 31: 235–240.
8. Umashankar T, Prasad T, Rajeshwari SH. Naevus lipomatosus superficialis: Clinicopathologic study of a case. Indian J Pathol Microbiol 2003;46:444-5.
9. Tunc S., Sezgin B., Yilmaz G., Gocun P. U., Kucuker I. Compression neuropathy caused by an unusual lesion: nevus lipomatosus cutaneus superficialis. Plast Reconstr Surg. 2011;127:72–74.

10. Cardot-Leccia N., Italiano A., Monteil M.C., Basc E., Perrin C., Pedeutour F. Nevus lipomatosus superficialis: a case report with a 2p24 deletion. *Br J Dermatol.* 2007;156:380–381. doi: 10.1111/j.1365-2133.2006.07622.x
11. Fatah S., Ellis R., Seukeran D.C., Carmichael A.J. Successful CO2 laser treatment of naevus lipomatosus cutaneous superficialis. *Clin Exper Dermatol.* 2010;35:559–560. doi: 10.1111/j.1365-2230.2010.03794.x.
12. Goucha S, Khaled A, Zéglauoui F, Rammeh S, Zermani R, Fazaa B. Nevus lipomatosus cutaneous superficialis: Report of eight cases. *Dermatol Ther (Heidelb).* 2011 Dec;1(2):25-30. doi: 10.1007/s13555-011-0006-y.
13. Ganjoo, Shikhar; Sawhney, MPS; Sharma, Uma<sup>1</sup>. Pedunculated Nevus Lipomatosus Cutaneous Superficialis of the Scrotum Successfully Removed by Excision. *Indian Journal of Paediatric Dermatology* 19(2):p 148-150, Apr–Jun 2018. | DOI: 10.4103/ijpd.IJPD\_39\_17
14. Castagna RD, Benvegnú AM, Dallazem LND, Brutti CS. Topical corticosteroid therapy: a treatment option for nevus lipomatosus cutaneous superficialis? *An Bras Dermatol.* 2018 Jan-Feb;93(1):158-159. doi: 10.1590/abd1806-4841.20186986
15. Kim HS, Park YM, Kim HO, Lee JY. Intralesional phosphatidylcholine and sodium deoxycholate: a possible treatment option for nevus lipomatosus superficialis. *Pediatr Dermatol.* 2012 Jan-Feb;29(1):119-21. doi: 10.1111/j.1525-1470.2011.01413.x.

## **FIGURE LEGENDS**



Figure 1A: Pre-operative image of the pedunculated, solitary nodule measuring 40×40 mm with a prominent stalk measuring 10 mm long. The swelling was skin-colored, nodular in appearance, with no sinuses or ulceration.



Figure 1B: Excised swelling with the stalk

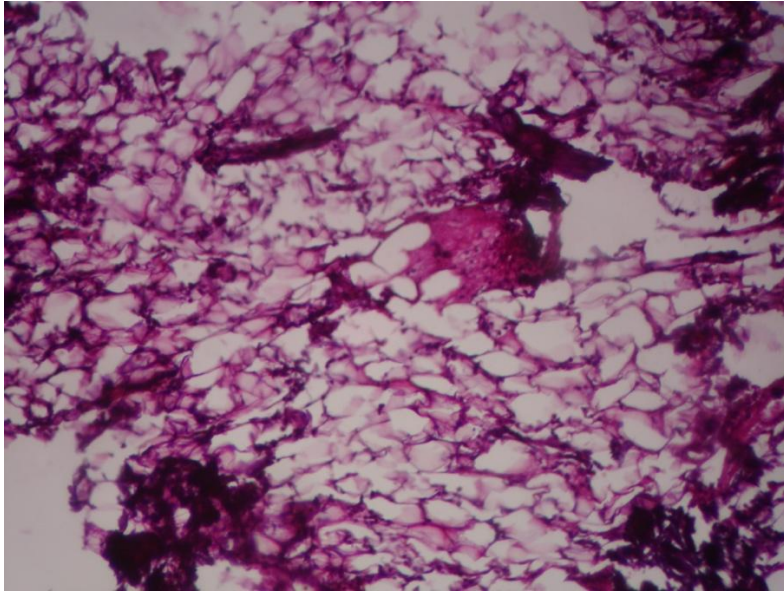


Fig 2A – Low power view showing hyperkeratosis, irregularly organized connective tissue and presence of mature adipocytes in the dermis.

UNDER PEER REVIEW